Risk factors for interstage death after stage 1 reconstruction of hypoplastic left heart syndrome and variants

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Objective: The risk of death during the interstage period remains high after stage 1 reconstruction for single ventricle lesions, despite improved surgical results. The purpose of this study is to identify risk factors for interstage death and to describe the events leading to interstage death.

Methods: A nested case–control study was conducted of 368 patients who underwent stage 1 reconstruction at a single center between January 1998 and April 2005.

Results: Among the 313 (85%) hospital survivors, there were 33 (10.5%) interstage deaths. Cases more frequently presented with intact or restrictive atrial septum (9 [27%] vs 4 [4%]; $P < .001$), were older at the time of surgery (5 [2–40] vs 3 [1–42] days; $P = .005$), had more postoperative arrhythmias (12 [36%] vs 15 [15%]; $P = .01$), and a higher incidence of airway or respiratory complications (12 [36%] vs 19 [19%]; $P = .04$). By multivariate analysis, only intact atrial septum (odds ratio 7.6; 95% confidence intervals 1.9–29.6; $P = .003$) and age at operation greater than 7 days (odds ratio 3.8; 95% confidence intervals 1.3–11.2; $P = .017$) were predictors of interstage death.

Conclusions: The presence of intact atrial septum and older age at the time of surgery are associated with a higher risk of interstage death. In addition, postoperative arrhythmia and airway complications are associated with a higher risk of interstage death in univariate analysis. The results of this study provide a focus for interstage monitoring and risk stratification of these high-risk infants, which may improve overall survival.

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Hypoplastic left heart syndrome (HLHS) and its variants present unique management challenges to the cardiac intensive care team after stage 1 reconstruction (S1R). Equally challenging is the prevention of interstage death (ISD), defined as death after hospital discharge and before admission for a planned stage 2 reconstruction (S2R). Hospital survival after S1R has improved as a result of continued innovation and refinement in surgical technique, earlier diagnosis and referral to tertiary care centers, and advances in perioperative care.[1-10] As a result, this group of congenital lesions once considered universally fatal in infancy now has a survival after S1R of 77% to 93%.[11-16] However, at most centers ISD rates are unchanged from historical rates of 7% to 15%.[17-22] Therefore, interstage mortality constitutes an increasing percentage of overall mortality in the current era and is attracting increased clinical and research attention.

The reported causes of ISD range from simple intercurrent illnesses to catastrophic shunt thromboses.[19,23] In attempts to identify those patients at risk for ISD events, past investigators have identified residual anatomic lesions, depressed myocardial function, elevated systemic vascular resistance, arrhythmia, and noncardiac factors such as seizures and feeding dysfunction as significant risk factors.[3,12,19-25] It is clear that after

Abbreviations and Acronyms

CI	= confidence interval
ISD	= interstage death
OR	= odds ratio
S1R, S2R	= stage 1 reconstruction, stage 2 reconstruction

S1R, many patients with single ventricle physiology have fragile hemodynamics and at times do not tolerate even minor stressors. Therefore, management strategies have focused on creating a more stable circulation through surgical and medical advances as well as identifying and better monitoring those patients at high risk for ISD. The goal of this study is to identify those patients who are at high risk for ISD, with a secondary goal of describing the events leading to ISD.

Patients and Methods

Study Design

We performed a retrospective case–control study including all patients who underwent S1R at the Children's Hospital of Philadelphia from January 1998 to August 2005. The study was approved by the Institutional Review Board (IRB No. 2004-10-2890); informed consent was waived. Hospital medical records, the Cardiac Center database systems, and outpatient cardiology records were reviewed to identify ISD and control patients. For study patients, additional data reviewed included echocardiography reports, cardiac catheterization reports, and operative reports, including extramural reports when appropriate. Outpatient cardiologists were contacted for follow-up information when necessary. For each case, 3 controls were randomly chosen from the cohort of patients who had undergone surgery within the same calendar year as the case and had survived to S2R.

Definitions

Patients who died before hospital discharge are designated as hospital deaths. Hospital deaths include patients transferred to referring hospitals who died before discharge from those units. For those patients transferred to a referring hospital, the transfer date was used for calculating length of stay. Inoperable patients were those not referred for S2R within 1 year of S1R owing to unsuitability for further palliation. Transplanted patients were those who underwent S1R but subsequently underwent transplantation before S2R. Hospital deaths, inoperable patients, and transplanted patients were excluded from the control group.

Anatomy and Patient-related Factors

All patients underwent S1R for lesions consisting of a single functional ventricle with obstruction to systemic outflow (Table 1). Anatomic diagnoses were established from echocardiography reports, cardiac catheterization reports, surgical notes, and autopsy reports. In cases of a disputed diagnosis, operative reports and autopsy reports were given precedence. The diagnosis of intact atrial septum was based on echocardiography or catheterization findings of absent atrial communication. Highly restrictive atrial septum was defined as an atrial septal defect measuring less than 2 mm and a restrictive Doppler flow pattern.[26]

TABLE 1. Preoperative anatomy: Subclass and variants

Category	Cases (n = 33)	Controls (n = 99)	*P*
HLHS (all)	25 (76%)	64 (65%)	.20
With aortic atresia	17 (52%)	44 (44%)	.48
HLHS variants			
Complex double-outlet right ventricle	0	9 (9%)	.11
Unbalanced atrioventricular canal	5 (15%)	9 (9%)	.33
Other*	3 (9%)	17 (17%)	.26

* *Other* includes double-inlet left ventricle with transposition of the great arteries (TGA) and systemic outflow obstruction, tricuspid atresia with TGA and systemic outflow obstruction, TGA with inadequate systemic ventricle and systemic outflow obstruction, and critical aortic stenosis/aortic hypoplasia with inadequate systemic ventricle.

Operative Technique

The standard S1R performed at our institution during the study period consisted of an atrial septectomy, division of the main pulmonary artery with side-to-side anastomosis to the ascending aorta, or homograft patch augmentation of the ascending aorta and aortic arch, with either a modified Blalock–Taussig shunt or a right ventricular–pulmonary artery conduit as the source of pulmonary blood flow.[12] From 1998 to 2002, the modified Blalock–Taussig shunt was used exclusively, whereas from 2002 to 2005 there was contemporary use of both shunt types. Five surgeons performed S1R during this period. Repair of additional intracardiac lesions was undertaken simultaneously where appropriate. Modified ultrafiltration was routinely used. In the postoperative period, neither delayed sternal closure nor extracorporeal membrane oxygenation was routinely used. The usual postoperative care during this time period included infusions of milrinone, dopamine, and fentanyl, normal ventilation, normothermia, and discontinuation of neuromuscular blockade the first postoperative night.

Data Analysis

Demographic data, operative factors, and postoperative events were recorded. Continuous variables are presented as mean (± standard deviation) for normally distributed data and median (range) for non-normally distributed data. Dichotomous variables are presented as count and percent. Categorical data were compared by the Fisher exact test or χ^2 test where appropriate. For comparison of continuous variables between cases and controls, a Student *t* test was used for normally distributed data and a Wilcoxon rank sum test otherwise. An a priori significance level of $\alpha = .05$ was used. Stepwise logistic regression using backward selection was performed to identify independent risk factors found to be significant from univariate analysis. Analysis was performed with STATA 8.0 (STATA Corporation 2003, College Station, Tex).

Main Results

During the study period, there were 33 (10.5%) ISDs (Figure E1). Of the original cohort of 368 patients, 263 (71%) went on to S2R, from which the control group was taken.

Risk Factors for ISD

Preoperative demographic and cardiac features analyzed are shown in Table 2. Patients with an intact or highly restrictive atrial septum

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TABLE 2. Preoperative variables: Demographics and cardiac features

Variable	Cases (n = 33)	Controls (n = 99)	P
Gender (% male)	19 (57%)	66 (67%)	.35
Birth weight (kg)	3.0 (1.5–4.2)	3.2 (1.6–4.5)	.43
Gestational age (wk)	40 (30–41)	39 (31–41)	.28
Age at time of surgery (d)	5 (2–40)	3 (1–42)	.005
Age at surgery > 7 d	10 (30%)	10 (10%)	.006
Birth weight < 2.5 kg	6 (18%)	15 (15%)	.68
Born remote to surgical center	14 (42%)	50 (50%)	.42
Prenatal diagnosis	15 (45%)	61 (62%)	.11
Syndromes and major malformations			
Major noncardiac anomaly	7 (21%)	16 (16%)	.5
Heterotaxy syndrome	3 (9%)	4 (4%)	.37
Cardiac variables			
Single left ventricle	0	14 (14%)	.02
Intact or highly restrictive atrial septum	9 (27%)	4 (4%)	<.001
Preoperative AV valvular insufficiency*	7 (21%)	14 (14%)	.34

AV, Atrioventricular. *Preoperative AV valvular insufficiency rated as moderate or greater by echocardiography.

were at significantly higher risk of ISD (9 [27%] cases vs 4 [4%] controls; $P < .001$). A single morphologically left ventricle was protective (0 cases vs 14 [14%] controls; $P = .02$). No other preoperative anatomic or functional cardiac factor was found to be significant. Among the patient demographic preoperative variables analyzed, only age at the time of operation was found to be significant: 5 days (2–40) in cases versus 3 days (1–42) in controls ($P = .005$).

No difference was found in any operative factor analyzed (Table 3). Of the postoperative factors evaluated (Table 4), airway or respiratory anomalies or respiratory complications were more common in cases (12 cases [36%] vs 19 controls [19%]; $P = .04$), and postoperative arrhythmias necessitating an intervention were more frequently documented in cases (12 cases [36%] vs 15 controls [15%]; $P = .01$). Of the cases with a significant postoperative

TABLE 3. Operative variables

Variable	Cases (n = 33)	Controls (n = 99)	P
Operative times (mean ± SD)			
Cardiopulmonary bypass time (min)	96 ± 34	95 ± 26	.85
Circulatory arrest time (min)	44 ± 13	46 ± 16	.63
Shunt type: RV–PA	3 (9%)	14 (14%)	.45
Delayed sternal closure	6 (18%)	14 (14%)	.58
Postoperative ECMO	2 (6%)	1 (1%)	.15
Surgical revision*	1 (3%)	6 (6%)	.68

SD, Standard deviation *RV–PA,* right ventricle–pulmonary artery conduit; *ECMO,* extracorporeal membrane oxygenation. *Surgical revision,* *revision of S1R within the same admission.

TABLE 4. Postoperative variables

Variable	Cases (n = 33)	Controls (n = 99)	P
Postoperative arrhythmia	12 (36%)	15 (15%)	.01
Postoperative arrest*	2 (6%)	1 (1%)	.15
Airway and respiratory:			
Need for reintubation	10 (30%)	17 (17%)	.11
Vocal cord paresis	4 (12%)	4 (4%)	.11
Diaphragm paresis	0	3 (3%)	.57
Tracheostomy	2 (6%)	2 (2%)	.26
Any airway complication†	12 (36%)	19 (19%)	.04
Nasogastric feeding at discharge	14 (42%)	49 (49%)	.48
Surgical gastric tube	5 (15%)	7 (7%)	.16
Postoperative seizure	6 (18%)	9 (9%)	.15
Abnormal MRI	10 (30%)	22 (22%)	.35
ECHO measures at discharge			
Ventricular dysfunction‡	0	3 (3%)	.57
Residual coarctation	1 (3%)	3 (3%)	1
AV valvular insufficiency§	10 (30%)	20 (20%)	.23
Neoaortic insufficiency‖	4 (12%)	4 (4%)	.11
On oxygen at discharge	8 (24%)	15 (15%)	.23
Discharge on > 3 medications	20 (60%)	41 (41%)	.06

MRI, Magnetic resonance imaging; *ECHO,* echocardiographic; *AV,* atrioventricular. **Postoperative arrest* requiring cardiopulmonary resuscitation including chest compressions. †*Any airway complication*: need for reintubation *or* vocal cord paresis *or* diaphragm paresis *or* trachesostomy. ‡*Ventricular dysfunction* rated at moderate or greater. §*AV valvular insufficiency* rated at moderate or greater. ‖*Neoaortic insufficiency* rated mild or greater.

arrhythmia, 4 had a re-entrant supraventricular tachycardia, 3 junctional ectopic tachycardia, 2 ectopic atrial arrhythmias and frequent atrial ectopy, and 2 ventricular tachycardia. One patient had high-grade heart block that resolved before discharge. Cardiac status at the time of discharge was no different between the groups, as evaluated by echocardiographic features, need for oxygen, and number of medications.

Multivariate logistic regression revealed intact or highly restrictive atrial septum (odds ratio [OR] 7.6; 95% confidence intervals [CI] 1.9–29.6; $P = .003$) and age at operation greater than 7 days (OR 3.8; 95% CI 1.3–11.2; $P = .017$) to be independent risk factors for ISD.

Circumstances of ISD

The circumstances of ISD events are detailed in Table 5. Death occurred at a median of 64 (18–257) days of age. The median time from discharge to ISD was 44 (2–188) days. Fourteen (42%) ISD patients experienced sudden, unexpected deaths at home. Interestingly, 11 (33%) patients died during a subsequent hospitalization. Of these, 4 died after a procedure (Nissen fundoplication, gastrostomy tube manipulation, shunt revision, and cardiac catheterization). The remainder were hospitalized with a wide range of diagnoses, including respiratory insufficiency and/or infections (n = 4), severe cardiac dysfunction (n = 2), and feeding dysfunction (n = 1). In 8 patients, information regarding the ISD event was not obtained. Autopsy results were available for 8 patients (Table E1).

TABLE 5. Circumstances of interstage death events

Patient	Age at OR (days)	Age at ISD (days)	Circumstances of ISD	Known risk factors at discharge*
1	2	104	Unknown	None
2	15	31	Observed emesis and arrest in hospital; GER	VCP
3	4	18	Sudden death at home; cyanosis on O_2	None
4	5	165	Died after G-tube exchange; severe TR	IAS; arrhythmia; failed extubation
5	3	NA	Sudden death at home	IAS; arrhythmia
6	5	51	Sudden death at home	None
7	2	31	Respiratory arrest; readmitted for CRI	Tracheostomy; failed extubation
8	2	125	Unknown	None
9	12	23	Unknown	Arrhythmia
10	5	140	Readmitted with PHTN; DNR	Arrhythmia; failed extubation
11	2	49	ICU arrest; readmitted with severe dysfunction	None
12	3	115	Sudden death at home; severe TR	Arrhythmia; failed extubation
13	2	96	Sudden death at home; recent *C. diff* admission	VCP; failed extubation
14	3	73	Sudden death at home	Failed extubation
15	4	57	Sudden death at home	IAS; arrhythmia; failed extubation
16	7	59	ICU arrest; RSV; interim stent for IAS	None
17	10	48	ICU arrest: aspiration pneumonia	None
18	40	135	Sudden death at home; likely BTS occlusion	IAS; arrhythmia
19	5	62	Died due to complications from Nissen	None
20	6	120	Sudden death at home associated with fever	None
21	3	78	Died after shunt revision. Died during readmission for shunt revision	None
22	7	38	Unknown	None
23	4	27	Sudden death at home; declining cardiac function	Arrhythmia
24	22	257	Unknown	Arrhythmia; tracheostomy
25	22	104	Died after complications of catheterization	None
26	5	64	Unknown	IAS
27	8	120	Sudden death at home; diarrhea, dehydration	IAS; arrhythmia
28	13	142	Sudden death at home; progressive cyanosis	VCP
29	8	62	Unknown	None
30	12	NA	Sudden death at home	IAS; arrhythmia; failed extubation, VCP
31	4	71	Sudden death at home likely related to bacteremia	Arrhythmia; failed extubation
32	4	59	Unknown	IAS
33	4	142	Readmitted with URI; observed apnea in hospital	None

OR, Operation; *ISD,* interstage death; *GER,* gastroesophageal reflux disease; *VCP,* vocal cord paresis; *G-tube,* surgical gastrostomy feeding tube; *TR,* tricuspid regurgitation; *IAS,* intact or highly restrictive atrial septum; *CRI,* chronic respiratory insufficiency; *PHTN,* pulmonary hypertension; *DNR,* Do Not Resuscitate order; *C. diff, Clostridium difficile*; *BTS,* Blalock–Taussig shunt; *URI,* upper respiratory infection. **Risk factors* including: IAS; age at surgery > 7 days; arrhythmia; airway/respiratory complications (failed extubation, vocal cord paresis, diaphragm paresis, or tracheostomy). ICU = Intensive care unit; RSV = Respiratory syncytial virus.

One patient had a documented shunt thrombosis, and another had evidence of diffuse thrombosis and emboli, but no occlusive thrombus in the shunt was found at the time of autopsy. In 3 patients there was evidence of myocardial ischemia and infarction, 2 of whom had significant involvement of the tricuspid apparatus. One patient had polymicrobial bacteremia, possibly associated with bowel perforation and bacterial translocation. In 2 patients, the autopsy did not suggest any cause of death.

Discussion

As recent advances have led to improved operative outcomes in HLHS and its variants, we hope this study will advance the development of management strategies to improve interstage outcomes. In this cohort, restriction at the level of the atrial septum and delayed time of surgery were found to be independent risk factors for ISD. In addition, univariate analysis found the occurrence of a perioperative arrhythmia or an airway or respiratory complication more frequently in cases. These modifiable risk factors provide intriguing targets for future interventions in addition to guiding risk stratification for the development of interstage monitoring protocols.

Patients with an intact or highly restrictive atrial septum are at high risk for mortality at every stage of single ventricle palliation, including the interstage period. These patients have evidence of abnormal pulmonary vasculature from birth, with "arterialization" of the pulmonary veins and

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dilation of pulmonary lymphatics.[26] As a result, they are more likely to have elevated pulmonary arterial pressures and are theoretically more susceptible to perturbations of pulmonary and systemic vascular resistance associated with interstage stressors. Recent research efforts have focused on fetal intervention, using either early surgical or interventional catheterization techniques to relieve the obstruction before the development of pulmonary vascular disease.[27] Whether these interventions will positively affect outcomes remains to be seen. It is unclear whether the administration of supplemental oxygen or other medications targeting the pulmonary vascular bed in the interstage period would be beneficial in this select patient population.

Delayed surgery in single ventricle lesions may occur as the result of late presentation, prematurity or low birth weight, presentation with multisystem organ dysfunction, and in some centers after primary consideration for transplantation. Past studies have shown that older age at time of presentation and at time of operation results in worse outcomes in this population.[21,26,27] In this study, we found that patients older than 7 days at the time of operation were more likely to have ISD. In most cases there is no advantage in delaying surgery; however, individual patients have survived delayed palliation, and in specific circumstances it may be necessary to delay S1R.[6] It is likely that this variable represents a marker for level of illness at presentation in the current cohort: well patients with no organ dysfunction are more likely to go to the operating room promptly and have a favorable operative and postoperative course.

Our finding that perioperative arrhythmia is more commonly identified in ISD patients agrees with previous studies.[17,21] Of the 12 ISD patients identified with significant postoperative arrhythmia, 9 (27%) were discharged receiving a medication specifically to treat or prevent arrhythmia. Only 1 patient from the entire cohort had a documented arrhythmia at the time of ISD and received cardioversion in the field for wide complex tachycardia, without return of spontaneous circulation. This patient was not among those known to have a perioperative arrhythmia. Arrhythmia may explain a significant proportion of unexpected ISD but represents a difficult target for retrospective study. Arrhythmia as a cause of death cannot be diagnosed at autopsy, nor is rhythm at the time of arrest always possible to ascertain in the field. Kaltman and associates[28] prospectively compared indices of heart rate variability in infants who underwent single ventricle palliation to those with 2-ventricle repairs of various congenital lesions. The authors demonstrated a significant reduction in heart rate variability and a higher baseline heart rate during the vulnerable period for ISD in these patients.

The causes of death in this series were varied, and no previously unrecognized residual technical issues were identified on autopsy. In an autopsy series of 122 deaths after S1R from 1980 to 1995, Bartram, Grunenfelder, and Van Praagh[23] concluded that, in their series, most deaths were attributable to potentially correctable residual technical issues. Data from our institution and other large centers demonstrate that many of these technical issues have been eliminated or substantially improved on in the current era.[11-15,21] Therefore, causes of ISD that previously constituted a smaller proportion of total post-S1R mortality (eg, arrhythmia, shunt thrombosis, viral infections) have become more prevalent. Fenton and coworkers[19] described a series of patients with shunt-dependent pulmonary blood flow who had ISD and found that autopsy-proven shunt thrombosis occurred in 33% of these patients. Other retrospective clinical studies have found the rate of shunt thrombosis to be from 8% to 16%,[1,3,13] whereas in our experience only 6% of ISDs were associated with possible shunt thrombosis on autopsy. Interestingly, Fenton's group[19] found no difference in the incidence of ISD or shunt thrombosis in patients taking aspirin and those receiving no anticoagulation. The ideal anticoagulation strategy remains an important area of discussion at every stage of single ventricle palliation.

High systemic afterload may play a significant role in the pathogenesis of unexpected ISD in this population. Recent studies have highlighted the important role of afterload reduction in postoperative care and long-term management.[3,8,25] In addition, there is mounting evidence that autonomic function is altered after cardiac surgery,[28] which may further impair the ability of these patients to compensate for wide swings in systemic vascular resistance associated with intercurrent illnesses and other interstage stressors. In this cohort, patients with significant atrioventricular valvular regurgitation or ventricular dysfunction in the postoperative period were frequently prescribed captopril, but oral afterload reduction was not routine.

It is unlikely that a single surgical or medical intervention will significantly improve current ISD rates owing to the heterogeneous nature of ISD events. It may be more effective to focus our attention during the interstage period on identifying those most at risk, increasing and standardizing surveillance, and investigating subtle abnormalities earlier. In a study using an intensive multidisciplinary interstage monitoring program, Ghanayem and associates[20] demonstrated an improvement in the ISD rate from 15.8% to 0% during the study period. At the time of discharge, study patients were provided with a scale, pulse oximeter, and journal wherein weights and saturation levels were recorded daily by parents. Thirteen of 24 patients were found to have a decrease in saturation level or failure to gain weight; these patients underwent S2R at a median of 3.7 months versus 5.2 months for those with the usual interstage course. In our cohort, although a significant number of patients died after rehospitalization, the most common scenario was a sudden unexpected death at home. In roughly half of these cases, either progressive cyanosis or an identifiable preceding minor illness was found. These symptoms, though nonspecific, may be early signs of impending ISD and require greater attention.

Limitations

The main limitations of this study relate to its retrospective nature. Data surrounding ISD events and autopsy reports were limited or unavailable for some patients, especially those cared for outside the referral area. During the study period, evaluation of airway or respiratory complications in all patients was not standard. The decision to perform bronchoscopy was at the discretion of the clinician; in general, only those patients who were especially symptomatic were referred for evaluation. Therefore, potentially not all patients with vocal cord paralysis were identified, and it is possible we may have missed more subtle vocal cord disease. In addition, small numbers of patients with specific risk factors limit the predictive power of this analysis.

Conclusions

ISD remains a significant source of mortality in HLHS and its variants, occurring in 10.5% of patients surviving to hospital discharge. Patients with an intact or highly restrictive atrial septum and those undergoing S1R at greater than 7 days of life are at significantly higher risk for ISD. In addition, the presence of a postoperative arrhythmia or postoperative airway or respiratory complication is associated with a greater risk of ISD in univariate analysis. The causes of ISD are multiple, and it is unlikely that a single surgical or medical intervention will significantly improve the incidence of ISD. The results of this study provide a foundation for prospective interstage surveillance focused on minimizing ISD and improving overall outcomes in patients with single ventricle.

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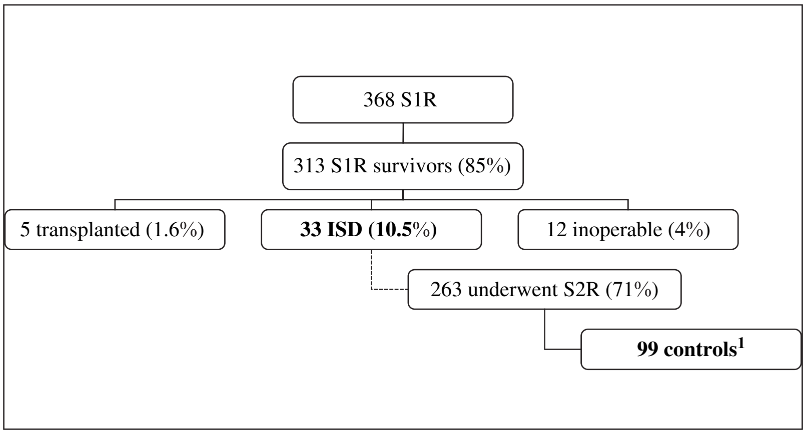

Figure E1. Patient population. Five patients were lost to follow-up after S1R; *S1R,* Stage 1 reconstruction; *ISD,* interstage death; *S2R,* stage 2 reconstruction.[1] Controls were randomly selected (3 for each case from within operative year of case) from the cohort of patients surviving to S2R.

CHD

TABLE E1. Results of available autopsies

Patient	Autopsy results
2	Old and new infarctions of the anterior papillary muscle of the tricuspid valve
11	Subendocardial and myocardial infarction and hemorrhage consistent with myocardial infarction
15	Nondiagnostic; right ventricular hypertrophy; small subdural hemorrhage
18	Shunt occlusion with septic thrombus
20	Severe cardiomegaly and right ventricular hypertrophy; residual coarctation 4 mm
21	Multiple systemic emboli and thromboses including pulmonary and renal
27	Hemorrhage and infarction of papillary muscle; residual coarctation 4 mm
31	Polymicrobial sepsis; possible bowel infarction

APPENDIX E1. Abbreviations and Acronyms

AA	Aortic atresia
AS	Aortic stenosis
AV	Atrioventricular
AVC	Atrioventricular canal
BTS	Blalock–Taussig shunt
C. diff	*Clostridium difficile*
CPB	Cardiopulmonary bypass
CRI	Chronic respiratory insufficiency
DHCA	Deep hypothermic circulatory arrest
DORV	Double-outlet right ventricle
DNR	Do Not Resuscitate order
DSC	Delayed sternal closure
ECMO	Extracorporeal membrane oxygenation
GER	Gastroesophageal reflux disease
GT	Surgical gastric feeding tube
HLHS	Hypoplastic left heart syndrome
IAS	Intact or highly restrictive atrial septum
ICU	Intensive Care Unit
IRB	Institutional review board
ISD	Interstage death
LV	Left ventricle
MA	Mitral atresia
mm	Millimeter
MRI	Magnetic resonance imaging
MS	Mitral stenosis
NG	Nasogastric feeding tube
PHTN	Pulmonary hypertension
RV–PA	Right ventricle to pulmonary artery shunt
S1R	Stage 1 reconstruction
S2R	Stage 2 reconstruction
TGA	Transposition of the great arteries
TR	Tricuspid regurgitation
URI	Upper respiratory infection
VCP	Vocal cord paresis